Recurrent Chylothorax Associated with Sarcoidosis

Robert J. Lengyel MD, Dean J. Shanley DO

Spontaneous chylothorax, an uncommon manifestation of pulmonary disease, has been described in association with infectious and inflammatory conditions involving the chest, thoracic trauma, and thoracic neoplasms. Isolated chylothorax is a rare manifestation of sarcoidosis and is usually associated with the presence of mediastinal and hilar adenopathy causing obstruction of the thoracic duct. We report a case of recurrent spontaneous chylothorax in a patient with parenchymal sarcoidosis without hilar or mediastinal adenopathy.

Chylothorax is usually the result of compromise of lymphatic flow through the thoracic duct. Neoplastic processes and trauma account for the majority of cases of chylothorax described in the literature.[1-2] Other etiologies include infectious and inflammatory diseases, congenital malformations of the lymphatics, and idiopathic causes. Sarcoidosis has rarely been reported as a cause of isolated chylothorax; three case reports in the literature describe patients with extensive intrathoracic adenopathy associated with sarcoidosis and complicated by chylothorax.[3-5] The following report describes a patient with parenchymal sarcoidosis only, complicated by recurrent right chylous effusions.

Case Report

A 42-year-old black man complained of increasing shortness of breath and right pleuritic chest pain during the past six months. The patient denied any history of recent or remote thoracic trauma. He was diagnosed with sarcoidosis six years prior and had experienced recurrent right-sided chylous effusions for more than two years despite corticosteroid therapy. Thoracentesis was performed monthly during the past year to relieve recurrent symptoms of shortness of breath and pleuritic pain.

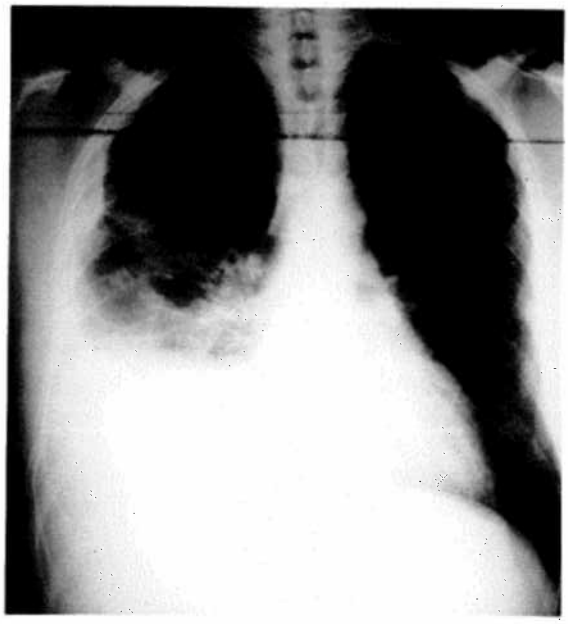

Fig 1.—42-year-old man with stage III sarcoidosis complicated by recurrent right chylothorax.

The admission chest radiograph (Fig 1) showed a large right pleural effusion and diffuse reticular nodular infiltrates involving both lungs. No evidence of hilar or mediastinal adenopathy was present. Findings were consistent with stage III (parenchymal) sarcoidosis. CT scan of the chest (Fig 2, 3) was obtained and confirmed the presence of a large right pleural effusion and bilateral reticular nodular infiltrates. The nodular opacities were small, innumerable, less than 1 mm in diameter and distributed diffusely throughout the parenchyma. Small densely calcified lymph nodes were identified in the mediastinum; no enlarged lymph nodes were present.

The patient opted for surgical intervention because of the chronic nature of the effusion and lack of efficacy of conservative therapy. The patient subsequently underwent right thoracotomy which revealed a large chylous effusion, a dilated thoracic duct, and periductal adhesions. Approximately 1.5 L of chylous fluid was removed from the right hemithorax. The thoracic duct was ligated and divided, followed by physical and chemical pleurodesis. The patient did well postoperatively and follow-up chest radiographs over a six-month period showed no recurrence of the chylous effusion.

Discussion

The thoracic duct normally ascends into the thorax through the aortic hiatus and maintains a position to the right of midline. The

Address reprint requests to:
Robert J. Lengyel MD
Department of the Army
Medical Department Activity
Kenner Army Community Hospital
Dept of Radiology
Ft Lee, Virginia 23801

Fig 2.—CT Scan of the chest shows a large right pleural effusion which proved to be chylous. Bilateral reticular nodular infiltrates are present throughout the lungs.

Fig 3.—CT scan of the chest using mediastinal (B) and lung (C) windows shows large right chylous effusion. Reticular nodular infiltrates are better demonstrated using lung windows. Small calcified lymph nodes are present, anterior to the right main-stem bronchus.

duct crosses obliquely to the left at approximately the level of the fourth or fifth thoracic vertebral body and continues to ascend into the left neck region for several centimeters. The duct then courses downward to join the venous system near the junction of the left internal jugular vein and subclavian vein. Obstruction or disruption of the thoracic duct anywhere along its course can lead to the development of chylothorax. Injury or obstruction to the lower portion of the duct typically results in a right chylous effusion, whereas involvement of the most cephalad portion of the duct usually results in a left chylothorax.[1]

Spontaneous chylothorax, although an uncommon manifestation of intrathoracic disease, has been reported in association with neoplastic processes, benign conditions, and trauma involving the chest.[1-2,6-7] Nix et al reviewed 302 cases of chylothorax and chylous ascites, of which 123 cases were of isolated chylothorax.[1] Of those cases of isolated chylothorax, approximately 55% of cases were traumatic in origin and almost 45% were spontaneous in nature. Thoracic surgery accounted for the majority of cases of chylothorax complicating *penetrating* trauma, and thoracic trauma without bony fracture resulted in the majority of cases associated with nonpenetrating trauma. Malignant neoplasms, especially lymphoma, resulting in extrinsic or intrinsic lymphatic obstruction account for the vast majority of spontaneous chylothoraces.[2] Benign lymphadenopathy is a less common cause of spontaneous chylothorax, contributing to less than 10% of the chylothoraces reviewed by Nix et al.[1]

Three previous cases of sarcoidosis with an associated chylothorax have been reported. Aberg et al reported extensive mediastinal and thoracic adenopathy causing obstruction of the thoracic duct, seen at postmortem examination.[3] Haigli and Keller reported mediastinal and hilar adenopathy, although the thoracic duct appeared unaffected at thoracoscopy.[4] The pathogenesis of the chylothorax was unclear and it gradually cleared after long-term corticosteroid treatment and pleural drainage. In a more recent case reported by Parker et al, a young woman with sarcoidosis developed a right chylous effusion and worsening mediastinal and hilar adenopathy within three months of fiberoptic bronchoscopy and transbronchial biopsy.[5] Thoracentesis and oral corticosteroid therapy resulted in resolution of the chylothorax.

Although spontaneous in nature, our patient's recurrent chylothorax was likely caused by the presence of periductal adhesions visualized during surgery, resulting in obstruction to the flow of lymph within the thoracic duct. Our patient had undergone transbronchial biopsy six years prior to the development of a persistent chylothorax, so there is no immediate temporal relationship between the effusion and the biopsy. Mediastinal and hilar adenopathy were present in our patient when the diagnosis of sarcoid was established six years prior; however, the adenopathy had regressed within two years. Imaging studies during the past four years showed no evidence of abnormally enlarged lymph nodes in the hilar regions or mediastinum, and the patient remained asymptomatic except for complaints of shortness of breath and pleuritic chest pain related to the recurrent chylothorax. During the progression of our patient's sarcoidosis fibrotic changes in the pleural space and perinodal regions probably contributed to the formation of periductal adhesions, gradual obstruction of the thoracic duct, and the development of recurrent chylous effusions. Unlike previous reports, this case is of interest because the recurrent chylothorax associated with sarcoidosis developed in the absence of lymphadenopathy, presumably caused by fibrotic changes in the pleural space and perinodal regions contributing to the formation of periductal adhesions and gradual obstruction of the thoracic duct.

References

1. Nix JT, Albert M, Dugas JE, Wendt DL. Chylothorax and chylous ascites: A study of 302 selected cases. *Am J Gastroenterol*. 1957; 28:40-55.
2. Bower GC. Chylothorax: observations in 20 cases. *Dis Chest*. 1964; 46: 464-468.
3. Aberg H, Bah M, Waters AW. Sarcoidosis complicated by chylothorax. *Minn Med*. 1966; 49:1065-70.
4. Haegli A, Keller R. Chylothorax associated with sarcoidosis. *Schweiz Med Wochenschr*. 1981;111: 125-8.
5. Parker JM, Torrington KG, Phillips YY. Sarcoidosis complicated by chylothorax. *So Med J*. 1994;87:860-62.
6. Cappell MS, Friedman D, Mikhail N. Chyloperitoneum associated with chronic severe sarcoidosis. *Am J Gastrolenterol*. 1993; 88:99-101.
7. Rockoff SD, Rohatgi PS. Unusual manifestations of thoracic sarcoidosis. *Am J Roentginol*. 1985; 144:513-528.